DIAGNOSIS

The differential diagnosis of the skin lesions was malignant syphilis (also known as lues maligna or ulceronodular syphilis), tertiary gummatous syphilis or an endemic treponematose. Histopathology showed an interstitial lymphohistiocytic infiltrate with formation of non-necrotizing granulomas. Giemsa, Ziehl-Neelsen and Grocott stainings were negative. The culture showed no growth of mycobacteria and fungi. During follow-up, the RPR increased to 1:256. The polymerase chain reaction (PCR) test for *Treponema pallidum* conducted on the skin biopsy was positive. A Treponema IgG immunoblot was performed and all bands were positive. Combined with the rise of the RPR titer, this confirmed the diagnosis of an active syphilis infection.

Malignant syphilis is a rare form of secondary syphilis often accompanied with the prodomal phase of fever, headache and myalgia.\(^1,2\) The first systematic studies of malignant syphilis were performed by Haslund\(^3\) and Neisser\(^4\), who differentiated malignant syphilis as a severe form of secondary syphilis from the gummas of tertiary syphilis. The following diagnostic criteria for malignant syphilis are defined as: strongly positive RPR titer, a severe Jarisch-Herxheimer reaction, characteristic macroscopic and microscopic morphology and rapid resolution of the lesions with antibiotics.\(^5\) The macroscopic characteristics are pleomorphic papulopustules, beginning ulcerations and deep ulcerations covered with crusts. The microscopic characteristics are an interstitial lymphohistiocytic and plasma cell-rich infiltrate with formation of non-necrotizing granulomas, sometimes in the presence of spirochetes. We diagnosed our patient with malignant syphilis because of the lamellar crusting, multiple ulcers, strongly positive RPR titer, a positive *treponema pallidum* PCR test of suspected syphilis lesions and rapid resolution of the lesions after antibiotic treatment.\(^7\)

Although tertiary gummatous syphilis was considered as a differential diagnosis, this was less likely in our patient. Lamellar crusting is not a feature of tertiary syphilis and gummatous disease generally takes years to decades to develop after initial infection, although progression may occur faster in HIV-positive patients.\(^8\)

In our patient, neurosyphilis was excluded with an ultrasound of the heart. In our patient, neurosyphilis was considered as a differential diagnosis, we consider this less likely in our patient. Lamellar crusting is not a feature of tertiary syphilis and gummatous disease generally takes years to decades to develop after initial infection, although progression may occur faster in HIV-positive patients.\(^8\)

Endemic treponematoses consisting of *Treponema pallidum* subsp *pertenue* (yaws), *T. pallidum* subsp *endemicum* (bejel), and *T. carateum* (pinta) were also considered as differential diagnoses. It is very difficult to distinguish venereal syphilis from the endemic treponematoses by serology only. However, yaws and bejel were less likely due to the patient’s country of origin and travel history. Pinta can be present in the Americas, including the Caribbean, but primary skin lesions due to pinta do not ulcerate. Furthermore, the rapid resolution of the lesions after treatment is not typical for pinta.

Penicillin is the best treatment for malignant syphilis: 10 to 14 days of IV treatment\(^9,10\), as well as 2.4 MIU intramuscularly, weekly for three weeks\(^11\), have shown good response. Malignant syphilis is considered as form of secondary syphilis, so with a confirmed infection within one year, a single dose of 2.4 MIU penicillin could also be considered. In our patient, neurosyphilis was not present, but the duration of infection and previous treatments were not documented. Therefore, we treated the patient with benzathin benzylpenicillin, 2.4 MIU intramuscularly for three consecutive weeks. The lesions disappeared completely in the following weeks leaving only hyperpigmentation.

In conclusion, atypical skin disorders in a patient with syphilis may be a form of malignant syphilis, especially in a HIV co-infected patient. The characteristic macroscopic and microscopic morphology, strongly positive RPR titer and rapid resolution of the lesions with antibiotics may lead to this rare diagnosis.

REFERENCES


